CASE 1997-10

Submitted by: Hans H. Goebel, M.D. and J.K. Mellies, M.D.
Dept. of Neuropathology, Mainz University Medical Center, Langenbeckstrasse 1, D-55131 Mainz /-Germany and Dept. of Neurology, Municipal Hospital,Osnabruck / Germany

Clinical History:

This 59-year-old woman was referred for further investigation of cognitive decline. She reported progressive forgetfulness especially with problems naming. She had started to make frequent notes, complained about poor concentration and being easily overcharged. Since her mid-thirties she had unilateral, throbbing headache of alternating sides about once a week. In recent years this headache has become rare. During the migraine attacks, numbness spreads from the right hand all over her right hemibody, and she has an expressive type of aphasia. Headache and neurologic deficits usually subside within half an hour. She experienced one similar event at the age of 19 years, but has been headache-free thereafter for almost 20 years. She reported urinary urge and incontinence for one year, which had not been alleviated by vesical neck surgery. There were no history of stroke or TIA, no mood disorder and no vascular risk factors, beside obesity and hyperlipidaemia.

Neurological exam was normal. Results of further investigation were as follows: The blood pressure was normotensive during 24-h monitoring. Echocardiogram and CW-Doppler ultrasound of the neck vessels were normal. By Holter-monitoring sinus rhythm, but no arrhythmias were encounterd. Electrocardiomyography showed sinus rhythm and a left anterior hemiblock. Chest X-rays were normal. Electroencephalography showed 9 Hz alpha activity with intermittent generalized 3-6 Hz slowing and left parieto-occipital intermittent rhythmic delta activity. Laboratory results were normal except for cholesterol of 255 mg/dl, triglycerides of 280 mg/dl, alkaline phosphatase of 217 U/L and (-glutamic transaminase of 37 U/L, reflecting steatosis and obesity. Homocysteine was 15.2 nmol/ml, fibrinogen 667 mg/dl. Vitamin B12, folic acid, HDL- and LDL-cholesterol, thyroid hormones, lues serology, antinuclear-antibodies, c-Anca, p-Anca, anti-phospholipid-antibodies, rheumatic factors, and C-reactive protein were within normal limits. Erythrocyte sedimentation rates were 13/31 over the first two hours. Cerebrospinal-fluid examination was normal.

The patient's sister, a niece, one daughter, and one son have the same disease, another daughter is genetically affected, but without clinical symptoms, and a third daughter has migraine with aura, but is genetically unaffected.

Material submitted: 1 plastic-embedded section, Richardson stain
1 electron micrograph

Points for discussion: 1. Diagnosis
2. Nature of the lesion